

Wilms' Tumours With Intracaval Involvement

Vicente Martínez-Ibáñez, MD, PhD, José Sánchez de Toledo, MD, PhD,
Marta De Diego, MD, Amparo Castellote, MD, Constantino Sábado, MD,
Germán Javier, MD, Alfredo Marqués, MD, and José Boix-Ochoa, MD, PhD

Since Gross established the basic rules for nephrectomy in Wilms' tumour (WT) in 1953, the management of nephroblastoma has been more straightforward. However, some cases with intravascular involvement, currently detected by ultrasound (US), may represent a daunting challenge for the surgeon. Inferior vena cava with tumour thrombus induced by WT can be asymptomatic and, if undetected, can contribute to poorer prognosis for two main reasons: possible neoplastic cells inside the thrombus and higher morbidity risk of surgery. From 1979 to 1993, 81 WT were studied by routine US. Intracaval thrombosis was diagnosed in four

(5%), in one of which the thrombus extended to the right atrium. In our experience, the surgical strategy in each of the four cases (100% survival) depended on the length of the thrombus and whether or not it infiltrated the vena cava wall. If the thrombus can be easily removed: complete resection. However, in cases of atrial thrombus, and more particularly if the thrombus involves the intima, we suggest the thrombus not be touched since the problem may be solved by preoperative and postoperative chemotherapy. Thus the favourable prognosis would be maintained and superfluous risky surgery avoided. © 1996 Wiley-Liss, Inc.

Key words: Wilms' tumour, intracaval thrombus, atrial thrombus

INTRODUCTION

Nephroblastoma is the most common renal malignancy in childhood [1]. Intracaval thrombus involvement (ITI) is an infrequent complication occurring in 4–10% of patients with these tumours [1,2]. The cardiac cavities (right atrium) are rarely involved (0.7–3%) [2,3]. In past reports [4], previously undiagnosed ITI was discovered during surgical resection. Most surgeons felt obliged to remove the thrombus, which resulted in higher complication rates [1–3].

Wilms' tumour has traditionally been approached by *preoperative* chemotherapy in SIOP protocols [1,6] or by primary surgical resection in the majority of other protocols [7,8]. In most cases, surgery of atrial thrombosis often requires sternotomy, abdominal laparotomy, and cardiopulmonary bypass (CPB) [3–5,9]. This approach was believed to be the safest and most efficacious treatment, although morbidity rates exceed 50% (43–73%) [6]. Massive haemorrhage was the most common complication followed by tumour embolization with acute cardiac decompensation or arrest [6].

Here, we add our experience in patients with ITI, particularly those with atrium involvement, to some previously described cases with good results after preoperative chemotherapy.

PATIENT DATA AND RESULTS

From 1979, when ultrasound (US) was used routinely at our centre, up to 1993, 81 Wilms' tumours were studied. Intracaval thrombosis was diagnosed in four (5%), in one of which the thrombus extended to the right atrium and presented one node located in the right pulmonary lobe.

Three patients presented ITI at infrahepatic level [4], and thrombi were removed at the time of ureteronephrectomy. No complications occurred as the inferior vena cava (IVC) was controlled above and below the thrombus, which centrally occupied the vascular lumen. All these patients were classified as stage II [10] and currently have no evidence of disease 10, 9, and 8 years, respectively, after surgery.

From the Hospital Infantil Valle Hebrón, Departments of Paediatric Surgery (V.M.-I., M.D.D., A.M., J.B.-O.), Paediatric Radiology (A.C.), and Paediatric Oncology Unit (J.S.d.T.), Barcelona; Hospital Germans Trias, Department of Paediatrics (C.S., G.J.), Badalona, Spain.

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Address reprint requests to V. Martínez-Ibáñez, M.D., Manuel Girona 86, 7-2d, 08034 Barcelona, Spain.

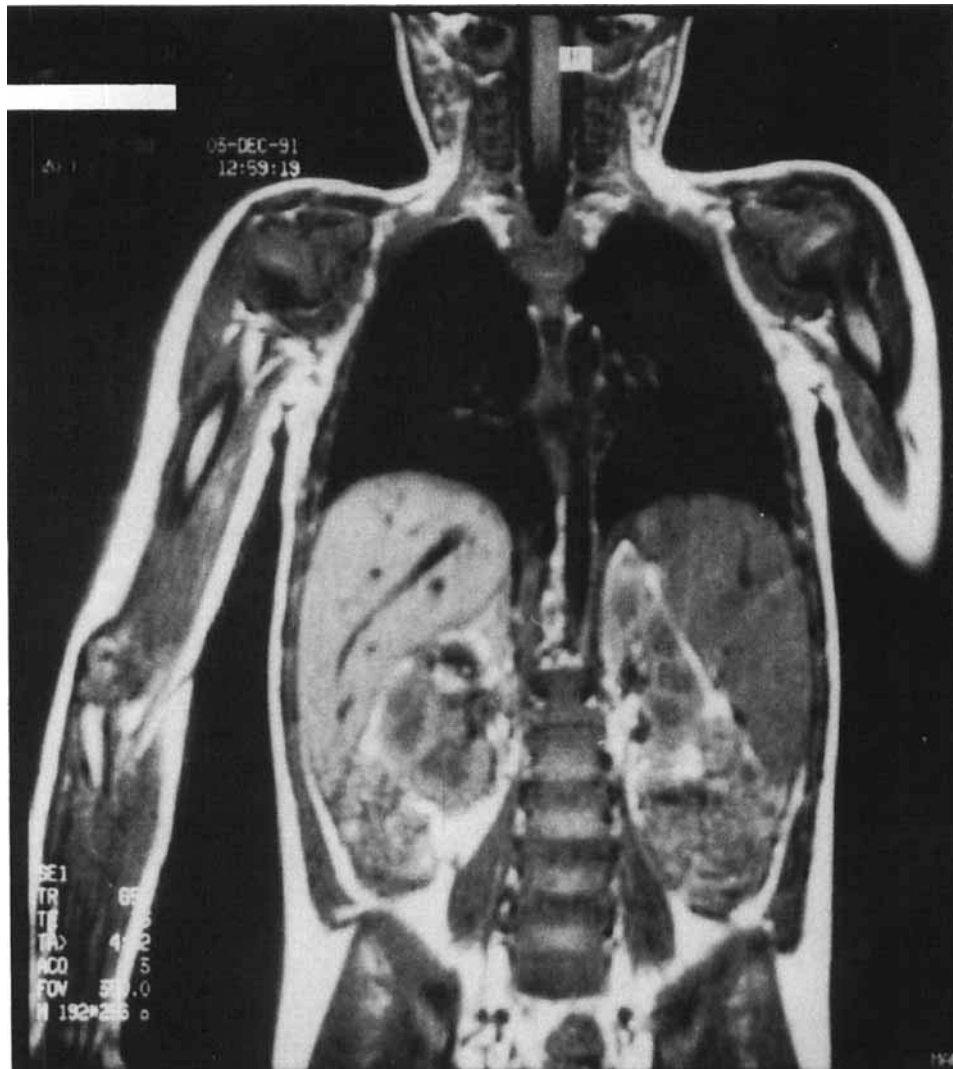


Fig. 1. MRI (coronal T1w TR/TE 658/15 nex:3) showing inferior vena cava thrombosis after preoperative chemotherapy.

A fourth case presented an intracardiac thrombus from the right Wilms' with probable pulmonary metastasis. In this patient, we performed sternotomy and abdominal midline laparotomy, raising the right side of the liver to expose the length of the IVC. The superior vena cava, right atrium, IVC below the thrombus, right and left renal veins, hepatic hilum, and hepatic veins were controlled by tourniquets. We opened the right renal vein and observed vascular wall involvement. As the vascular intima was invaded, we then attempted carefully to dissect the vessel-thrombus interface as in endarterectomy, but this proved impossible and an intracaval thrombus biopsy, right ureteronephrectomy-adrenalectomy was performed, once the metastatic pulmonary node had been removed at the same surgery. Following the SIOP protocol, this patient received preoperative chemotherapy and underwent

postoperative treatment as stage IV-A. The intracaval thrombus biopsy presented no viable tumour cells. Currently, 3 years later, the patient continues in good health with no evidence of disease.

Diagnostic imaging was performed with real-time ultrasound (US), a useful technique for diagnosing these tumours and following the response to treatment. CT scan was used for detecting possible metastases and MRI to control the evolution of the fourth patient.

DISCUSSION

Caval invasion shows no specific symptoms and is often asymptomatic [2]. For this reason, US [4,6] should be performed routinely to control possible thrombi and if these are present, to screen vascular extension. Right



Fig. 2. MRI (coronal T1w TR/TE 500/17 nex:4) showing inferior vena cava recanalization after postoperative chemotherapy.

Wilms' tumour appears to have more tendency to provoke thrombi than the left since the vein is shorter [2]. However, the left kidney has more possibilities of draining venous supply through the adrenal, lumbar, and gonadal veins [2,5].

Although recommended in previous studies, urography, cavography, and cardiac catheterization were not used [4,6]. In our opinion, real-time US seems to be the best imaging procedure for ITI. US is noninvasive and has been reliable in predicting both the presence and the level, as well as in following the response of a tumour to preoperative chemotherapy. Intraoperative US may help

to define the thrombus level. CT scan is useful in the assessment of metastases and may add information on intravascular disease, but it is insufficiently sensitive in locating total extension. MRI is of value for controlling intracaval thrombus evolution. Its coronal T1 weighted (w) images offer the surgeon anatomical relationships, whereas axial gradient-echo (gre) and T2w images clearly display the vena cava lumen, the thrombus within, and collateral vein dilatation. One must strive to obtain the necessary information at the lowest cost and avoid unnecessary exposure to radiation. Imaging protocols must be made as rational as possible and be based on

the best available current evidence. Angiography, which is invasive and expensive, cannot reveal the pathologic nature of the tumour with certainty. It does not change prognosis and may miss a second tumour [6].

In these cases, surgical strategy is based on thrombus location, either infrahepatic, retrohepatic, or atrial (4) and on whether the thrombus adheres to the vascular wall.

Thrombi at the infrahepatic level require control of the IVC above and below the thrombus with additional control of the contralateral kidney vein. In this case, uretero-nephrectomy can be performed easily and the thrombus removed at the same time.

If the thrombus is located at the retrohepatic level, the same vascular vessels as before and the hepatic hilum should be controlled. We recommend that the liver, particularly the right side, should be freed.

If the thrombus is located in the right atrium, we recommend control of the superior vena cava, right atrium, and hepatic veins. When the IVC is opened, the thrombus can be totally removed. At this moment, real-time US may be highly useful to ascertain the peak of the thrombi.

When a thrombus adheres tightly to the endothelium or infiltrates the vascular wall, our experience with our fourth patient taught us that the best way to proceed may be not to proceed and leave the thrombus alone. Prior to preoperative chemotherapy, our patient presented ascites, right pleural effusion, and intra-atrial thrombus by real-time echocardiography and total IVC obstruction with consequent azygos collateral channel opening by MRI. After three doses of preoperative chemotherapy [11] with epirubicin, vincristine, and actinomycin D at another centre, control CT scan showed that the Wilms' tumour had decreased and the ascites and pleural effusion had disappeared together with the presence of a right, presumably metastatic, pulmonary node. MRI revealed persistent IVC thrombus up to right atrium level although the intra-atrial thrombus had disappeared (Fig. 1). Biopsy taken at surgery from the intracaval thrombus contained no tumoural cells. Following postoperative chemotherapy (SIOP 9), real-time US and, particularly, MRI showed IVC recanalization with stenosis maintaining collateral channels open (Fig. 2). This patient was not given anticoagulant therapy, and, therefore, it must be supposed that the thrombus decrease and the lack of tumoural cells in the thrombus were due solely to preoperative chemotherapy.

Despite the fact that few cases of atrial and intracaval thrombus involvement have been reported in the literature, it seems clear that the favourable prognosis depends basically on tumour stage and histologic type. Possible complications would then result from aggressive surgery with manipulation of the thrombus, particularly when it has reached the atrium or adheres to the vascular wall.

In conclusion, if confluent veins can be well controlled

and the intracaval thrombus easily removed, we recommend complete resection. However, except for very rare cases of intracardiac thrombus with life-threatening symptoms urgently requiring surgery with CPB, in cases of atrial thrombus and particularly if the thrombus involves the intima, before exposing the patient to CPB or other aggressive surgery, we suggest the thrombus not be manipulated since it may be resolved by preoperative and postoperative chemotherapy. Nevertheless, if chemotherapy fails, surgery should be performed. The favourable prognosis would be maintained and superfluous risk surgery avoided.

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